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Didelphic Uterus and Obstructive Hemivagina with Ipsilateral Renal Agenesis Complicated by Pyocolpos

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The true incidence of reproductive tract anomalies in women is unknown, but has been reported as ranging between 0.1% and 3.8% [1]. Variation may be due to inaccurate diagnosis or defects in women with no symptoms that go undetected throughout their lifetime. Many factors contribute to the development of anomalies during the first trimester of gestation, such as exposure to extra- or intrauterine radiation, infection or teratogenic drugs (e.g. thalidomide and diethylstilbestrol), and anomalies may be due to polygenetic and familial inheritance as well [2,3].

Didelphic uterus with renal agenesis and obstructive hemivagina is a malformation of the genitourinary system occurring from embryologic arrest during the eighth week of pregnancy that affects the adjacent müllerian and metanephric ducts simultaneously [4]. It consists of defective lateral fusion of the caudal portions of the müllerian ducts during weeks 8–12 of pregnancy and also includes abnormalities linked to non-absorption of the septum at 20 weeks of gestation. Unilateral renal agenesis occurs when one or both ureteral buds fail to form or degenerate, and the metanephric blastema therefore does not differentiate into nephrons. Consequently, didelphic uterus is usually associated with a varying degree of vaginal septum and an absent, hypoplastic or duplicated kidney on the side with the obstruction [5,6]. The obstructive structure causes symptoms and signs to manifest at menarche, such as cyclic pelvic pain, fever, and abscess formation secondary to hematocolpos, hydrocolpos or pyocolpos [5,6].

In 1988, the American Society for Reproductive Medicine developed a classification system that allows anomalies to be organized according to major uterine anatomic types [7]. According to this system, our case was categorized as a class III uterine anomaly and class IIa vaginal anomaly. The workup, operative findings and therapeutic management of the condition will be described.

A 26-year-old unmarried Filipino woman was referred by a local clinic because of increasingly foul-smelling vaginal discharge over several months. She stated that her menarche occurred at age 13 with normal pubertal events, but her menses were irregular and characterized by hypomenorrhea and dysmenorrhea. During the few months prior to her referral, she began feeling severe lower abdominal pain particularly during her menstrual period, accompanied by hypomenorrhea with dark brownish spotting, followed by a foul-smelling vaginal discharge. The patient visited a local clinic where, on suspicion of pelvic inflammatory disease, medical treatment was given in vain. Then, she was referred to our outpatient department. A pelvic examination revealed a bulging mass over the left vaginal wall. An intravenous pyelogram was ordered to evaluate renal system function, with the finding of left kidney agenesis (Figure 1). Renal function was normal. Pelvic ultrasound demonstrated a double uterus (Figure 2A) and a homogeneous echolucent mass under the left cervix (Figure 2B). Computed tomography subsequently revealed the urinary system as having a solitary normal right kidney, with absence of a left kidney (Figure 3A), a didelphic uterus (Figure 3B), dilation of the left cervical canal, and vaginal fluid collection (Figures 3C and 3D), suggesting hydrocolpos, hematocolpos or pyocolpos. A relief operation was subsequently arranged. Examination was performed under anesthesia and the left lateral vaginal mass was palpated. A small opening was noted over the left lateral vaginal wall from which pus-like fluid was draining out. Hysteroscopy revealed no communication between the duplicated endometrial cavities, and a single ostium was visible from both uteri individually. A longitudinal vaginal septum was excised using electrocautery. Fine absorbable sutures were used to re-approximate the vaginal mucosa and prevent

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bleeding. Both cervices were then visualized. The patient recovered without complications. Two weeks later, postoperative pelvic examination revealed a healed vaginal mucosa and two cervices visible at the vaginal apex, indicating a good postoperative prognosis.

Didelphic uterus with unilateral obstructive hemivagina is the least common anomaly of the müllerian ducts but has the best prognosis in terms of alleviation of symptoms and future fertility [2,4]. The clinical symptoms of didelphic uterus usually manifest at menarche, including dysmenorrhea, hypomenorrhea, dyspareunia, and the presence of an intra-abdominal or pelvic mass, with infertility problems in the adulthood [5]. Early diagnosis is important in young girls who complain of obstructive symptoms, so that proper treatment can be chosen. But, because it is not a common anomaly, clinicians often misdiagnose the condition. For one thing, the menstrual period from the non-obstructive side is regular, delaying diagnosis of outflow obstruction that may cause amenorrhea and cyclic pain. Moreover, when symptoms of cyclic dysmenorrhea bring these girls to visit their physicians, they are usually given hormone treatment for having diminished or eliminated menses, inadvertently causing a delay in diagnosis [8]. Furthermore, the lower abdominal pain may be mistaken for pelvic inflammatory disease and treated with anti-inflammatory drugs.

Figure 1. Intravenous pyelogram: tiny opacity over right renal shadow of suspected artifact; non-opacification of left collecting system, with suspected left collecting system agenesis.

Prompt diagnosis and excision of the obstructed vaginal septum can relieve these symptoms completely and prevent further sequelae [9,10]. Diagnostic methods include hysterosalpingography, transvaginal ultrasound, computed tomography, magnetic resonance imaging, laparoscopy, and hysteroscopy. Hysteroscopy can detect intrauterine adhesions and communication between the duplicated endometrial cavities. Magnetic resonance imaging has 96–100% accuracy in classifying uterine anomalies, while transvaginal ultrasound has 85–92%, and hysterosalpingography has 6–55% [11].

Women with non-obstructive müllerian duct anomalies, such as didelphic uterus and unicornuate uterus, usually do not need surgical correction. Surgery is to preserve fertility and relieve symptoms; for these purposes, complete surgical excision of the obstructing vaginal septum is recommended. Precautions must be taken not to enter the bladder or rectum during the dissection. A Foley catheter placed in the bladder and a finger placed in the rectum will help define the anatomy.

Nevertheless, fertility in women with didelphic uterus is notably impaired. Some studies performed to

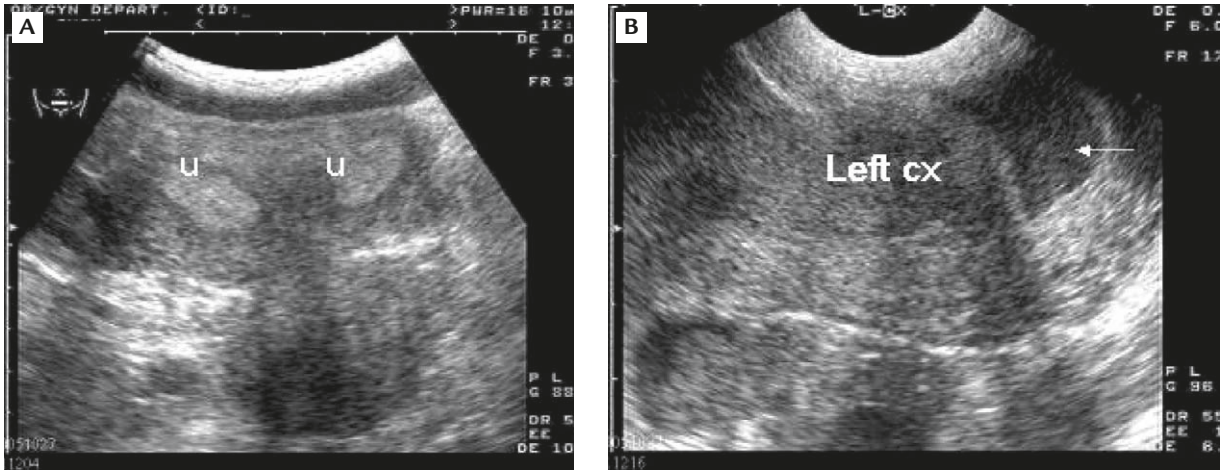

Figure 2. Pelvic ultrasonography: (A) external fundal cleft with wide divergence of endometrial cavities; (B) left cervix content with echolucent density (arrow). u = uterus; cx = cervix.

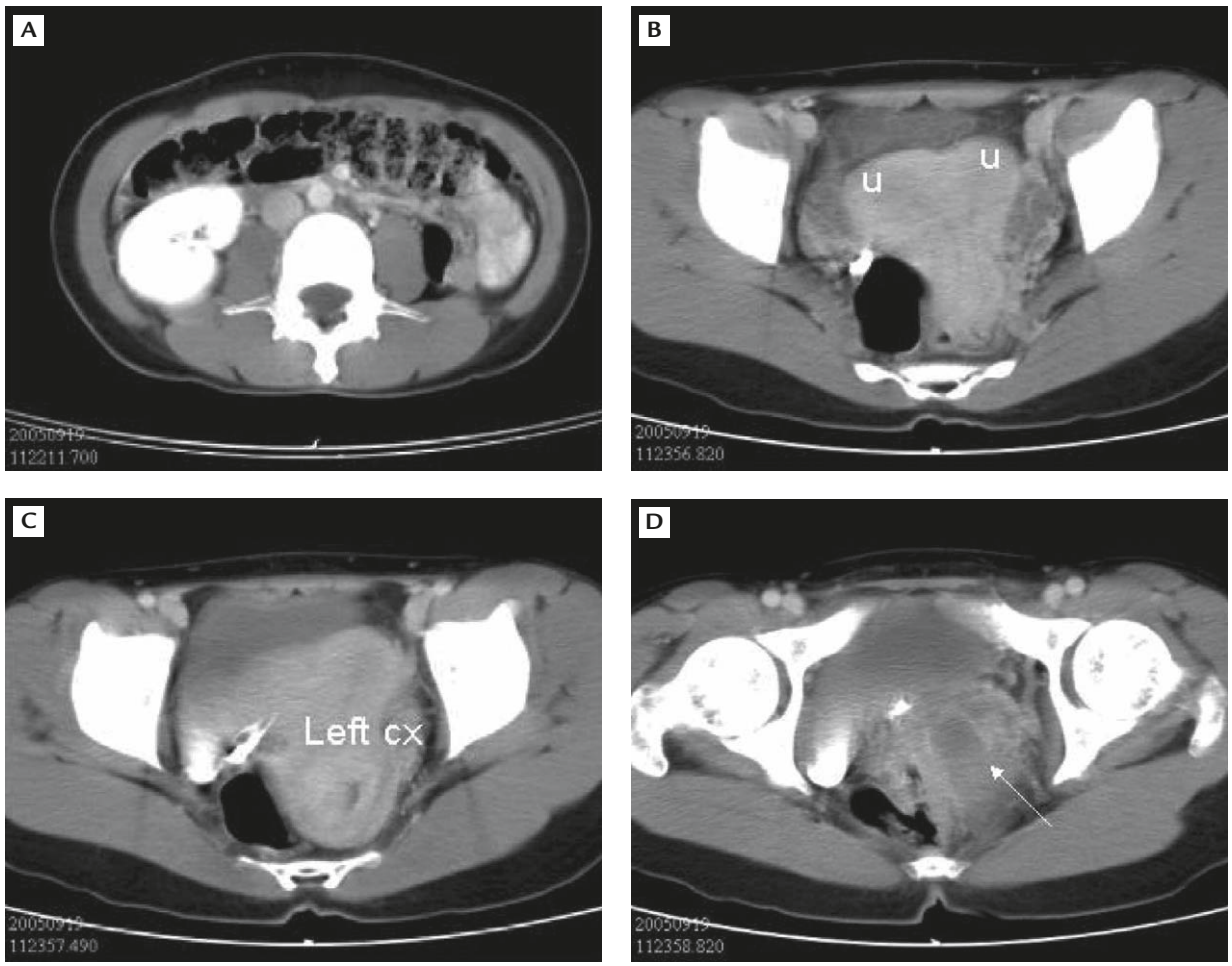

Figure 3. Computed tomography: (A) absence of left kidney is noted; (B) two uterine horns are widely separated and two cervical canals are noted; (C) dilatation of left cervical canal and vagina with fluid collection; (D) dilatation of left vagina with fluid collection (arrow). u = uterus; cx = cervix.

evaluate the reproduction of women with uterine anomalies found that successful pregnancy rates ranged from 37% to 40% in cases with didelphic uterus, similar to those for unicornuate uterus [9]. Endometriosis was well documented in these patients and might also contribute to a low pregnancy rate [12]. Another study reported 70% successful pregnancy outcomes, with preterm delivery at 20%, fetal growth restriction among 10%, and breech presentation in 43% of such pregnancies. The cesarean delivery rate was 82% [6].

In conclusion, obstructive müllerian anomalies are unusual and most often become apparent in childhood and adolescence. When patients begin to have obstructive symptoms, they are often misdiagnosed. In our case, the symptom of cyclic lower abdominal pain was noted in adolescence, and the patient took painkillers to relieve dysmenorrhea. Later, she suffered from purulent vaginal discharge and leukocytosis, leading to a presumptive diagnosis of pelvic inflammatory disease. Consequently, anti-inflammatory drugs were given for treatment at a local hospital. It seems possible that a small opening in the obstructive longitudinal vaginal septum allowed migration of pathogenic bacteria.

Our case is worth reporting because it underlines the importance of doing a thorough pelvic examination. A rare anomaly could be missed by a physician who fails to examine the female genital tract carefully. When abnormal bleeding, amenorrhea or cyclic pelvic pain are present in adolescents, clinicians must consider müllerian anomalies during differential diagnosis. The diagnostic methods described above are useful for delineating the anatomy. A prompt, accurate diagnosis is crucial, and the goals of therapy for young women with müllerian duct anomalies are relief of obstructive symptoms, restoration of normal menstrual egress and sexual function, and preservation of future fertility. Treatment is therefore unnecessary for patients without obstructive symptoms. We emphasize the need for clinical physicians to be familiar with the anatomy and physiology of these disorders in order to protect the patient from diagnostic delays and developing complications.

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